

**AB0164 DISEASE PROGRESSION IN 282 PATIENTS WITH  
UNDIFFERENTIATED SSC – DATA FROM THE GERMAN  
NETWORK FOR SYSTEMIC SCLERODERMA**

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**Background:** Systemic sclerosis (SSc) is a heterogeneous multisystem connective tissue disease, usually subdivided into the main SSc subsets, e.g. limited (lcSSc), diffuse SSc (dcSSc) and SSc overlap syndromes. However, some patients present with symptoms suggestive of, but not conclusive for a diagnosis of definite SSc. This subset has been referred to as undifferentiated or very early SSc. It was defined as positive RP together with at least one further feature of SSc and/or detectable SSc-specific autoantibodies.

**Objectives:**

**Methods:** Up to date, more than 3400 patients have been registered within the German network for systemic scleroderma. Disease progress after initial patient registration and further follow-up visits was analysed to determine, whether clinical features in patients with undifferentiated SSc change over time into definite SSc.

**Results:** Among 3473 registered patients, 8.1% (282/3473) were diagnosed with undifferentiated SSc. Of these, 87.5% were female with a mean age at onset of 59.9±1.5years. A significant difference was detectable comparing patients with undifferentiated SSc and lcSSc (54.8±0.5years; p<0.001) as well as dcSSc (49.8±0.6years; p<0.001). Positive antinuclear antibodies (ANA) were detectable in 219 (77.7%) patients; of these 40.2% were anti-centromer (ACA) positive and 12.8% anti-topoisomerase antibody (ATA) positive. No substantial difference regarding organ manifestations between patients with or without existing ANAs was found.

34.0% of undifferentiated SSc patients suffered from sicca symptoms, followed by 31.2% with gastrointestinal (GI) involvement, 30.5% with musculoskeletal involvement, 14.5% with lung fibrosis, 6.0% with heart involvement, 5.0% with pulmonary arterial hypertension (PAH) and 0.4% with renal crisis. Musculoskeletal involvement, sicca symptoms and heart involvement were found in a similar percentage in patients with undifferentiated and lcSSc. Lung fibrosis, PAH and GI involvement occurred significantly less in patients with undifferentiated SSc compared to lcSSc. Within a mean follow-up time of 3.6years, the majority of the patients classified initially as undifferentiated SSc, remained within this subset (76.6%), while only 16.3% converted into lcSSc. 4.6% were classified as dcSSc and 3.2% as SSc overlap syndromes.

**Conclusions:** Patients classified as undifferentiated SSc, develop clinical features suggestive of SSc at a significantly older age than other subsets (i.e. 5–10 years). During follow up progression into a limited, diffuse or SSc overlap syndrome subset was observed, however, the majority of undifferentiated SSc patients remained stable and did not develop definite SSc.

**Disclosure of Interest:** None declared

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